

Recurrent papilloedema and early onset optic atrophy in Behçet's syndrome

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Abstract

Two patients with Behçet's syndrome and intracranial hypertension are reported. One developed a recurrence of papilloedema while receiving treatment but eventually made a full recovery, whereas the other developed optic atrophy within three months of onset despite treatment.

Behçet's syndrome is a chronic inflammatory disorder of unknown cause,¹ in which 10-25% of patients have neurological manifestations.² Several patterns of neurological illness have been described,³ which include a brain stem syndrome, a meningo-myelitic illness, and an organic confusional state. Intracranial hypertension associated with Behçet's syndrome was first described in 1959,⁴ but it has only recently become recognised as one of the features associated with nervous system involvement.⁵ Considerable uncertainty remains about its pathogenesis, natural history, and response to treatment. We describe two cases with quite different clinical outcomes.

Case reports

PATIENT NO 1

A 30 year old white man presented with a six month history of recurrent conjunctivitis and oral, scrotal, and perianal ulcers. Within two months he developed an arthritis of the left knee and ankle which responded to benorylate. Ten months later he developed a left ileofemoral vein thrombosis and was treated for eight months with aspirin 37.5 mg daily, dipyridamole 100 mg four times a day, and stanozolol 5 mg daily. Treatment was then continued with azathioprine 50 mg in the morning and prednisolone 5 mg. He remained well for the next 20 months apart from one further hospital admission because of a right femoropopliteal vein thrombosis. Six months after the second thrombosis he was readmitted with a 10 week history of intermittent headache. There were no focal neurological signs, the cranial nerves were intact, but there was bilateral papilloedema with enlarged blind spots. Routine haematological and biochemical investigations were normal. A computed tomographic (CT) scan with contrast showed that the ventricles were of normal size with no focal lesion and no evidence of dural sinus thrombosis. Examination of the cerebrospinal fluid was normal apart from a raised pressure of >300 mm of water. He was treated with five intravenous pulses of 1 g methylprednisolone followed by 60 mg prednisolone

and 150 mg azathioprine daily. Symptomatic improvement occurred within six days of the first pulse of methylprednisolone and the fundal appearances reverted to normal within one month. Prednisolone was reduced gradually but six months later, while receiving 20 mg daily, the headaches and bilateral papilloedema recurred. He again responded to two further pulses of methylprednisolone and increase of the prednisolone to 40 mg daily. Three years later he is asymptomatic, takes 5 mg prednisolone daily, and his fundi and visual acuity are normal.

PATIENT NO 2

A 36 year old white woman presented with recurrent painful mouth and vulval ulcers of six months' duration and one episode of swelling of the left knee. One year after the onset of these symptoms she developed diarrhoea and a foul smelling vaginal discharge. Barium enema showed a rectovaginal fistula, and a temporary transverse defunctioning colostomy was performed, which was reversed two months later when she was asymptomatic. She remained well for 18 months when she again presented with a four week history of intermittent frontal headaches and blurring of vision. She had normal visual acuity, enlargement of both blind spots with constriction of the peripheral visual fields, and a right sixth nerve palsy. Fundoscopy showed florid bilateral papilloedema with scattered haemorrhages (figure (a)). Routine haematological and biochemical investigations were normal. A CT scan with contrast showed small ventricles with no evidence of dural sinus thrombosis or a space occupying lesion. Examination of the cerebrospinal fluid was normal except for a raised pressure of >300 mm of water. She was treated with three intravenous pulses of 1 g methylprednisolone followed by oral prednisolone (40 mg/day) and azathioprine (50 mg twice daily). Twenty four hours after her first pulse of methylprednisolone her right sixth nerve palsy had resolved and within seven days her visual symptoms had improved. At follow up a month later the visual acuity remained normal, even though the peripheral visual fields were constricted inferiorly. The papilloedema and haemorrhages had resolved but bilateral optic atrophy was now evident (figure (b)).

Discussion

Both of our patients satisfy the criteria necessary for the diagnoses, of Behçet's syndrome⁶ and intracranial hypertension.^{7,8} Intracranial

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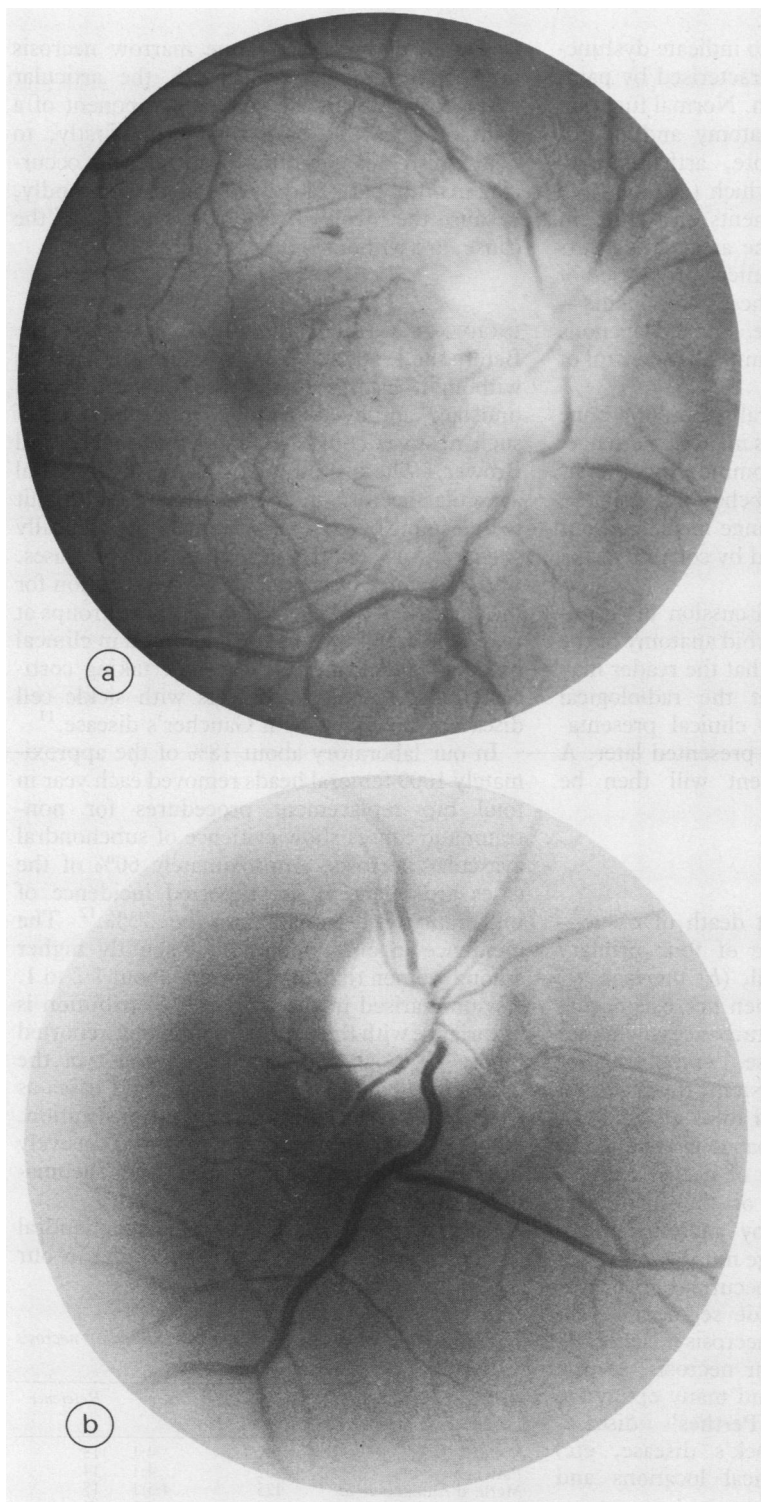
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hypertension may occur at any stage during the course of Behçet's syndrome⁹⁻¹³ and may be the presenting feature.^{9, 10} The natural history of this condition is not known. Spontaneous resolution of the papilloedema has been reported in a single case report,⁵ but all other reported patients had undergone treatment with steroids and some had also received immunosuppressive treatment. Some patients, with persistent symptoms and signs, needed repeated lumbar punctures^{9, 10, 13} or surgical treatment.^{9, 13} The eventual outcome is not known because the duration

of follow up in most reported cases was less than six months,^{5, 10, 13} but the prognosis seems to be good. Two patients were reported to have developed a recurrence of the papilloedema when the dose of steroids was reduced,^{10, 11} but the fundal changes responded to a subsequent increase in the dose. Only two patients developed optic atrophy¹³; at presentation they both had CT scans which showed the features of intracranial hypertension, and although both had angiography, venous sinus thrombosis could only be shown in one. Optic atrophy developed after one year and two years respectively.

Although sinus thrombosis has not been consistently proved to be responsible for the pathogenesis of intracranial hypertension in Behçet's syndrome it has been suggested that it is the underlying pathology.⁹ More recently, digital subtraction angiography has been suggested to be the radiological investigation of choice as it disclosed dural sinus thrombosis in two patients who had normal CT scans.¹¹ A larger study comparing the relative efficacy of these two procedures in diagnosing cranial venous sinus thrombosis showed, however, that in those patients in whom venous sinus thrombosis was shown by digital subtraction angiography only 10% had normal CT scans.¹⁴ Even though the clinical presentation, treatment, and prognosis of patients with or without proved intracranial thrombosis seems to be similar it remains unclear whether intracranial thrombosis is the only pathogenic process.

Intracranial, hypertension, with or without proved dural sinus thrombosis, is an uncommon complication of Behçet's syndrome and the optimal treatment and eventual outcome remain uncertain. We describe two patients who developed intracranial hypertension and papilloedema four years after the onset of Behçet's syndrome but whose outcome was quite different.



Fundal appearances (R) at presentation (a) showing papilloedema and haemorrhages and three months later (b) when the changes associated with optic atrophy are evident.

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